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Rhabdomyosarcoma of the kidney

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ABSTRACT

Rhabdomyosarcoma is considered the most common soft tissue sarcoma arising in patients younger than 15 years old, accounting for 5%–10% of childhood solid tumors. Sarcoma of the kidney represents 1% of all primary renal malignancies. Primary renal rhabdomyosarcoma is a very rare entity with limited number of cases reported in the literature. In this paper we present two cases of primary renal rhabdomyosarcoma in pediatric patients. The two tumors involved the renal parenchyma and occurred in 2-year-old girl and 6-year-old boy, respectively. Histopathology examination and immunohistochemistry studies confirm the diagnosis of embryonal rhabdomyosarcoma with pleomorphic component, and pleomorphic rhabdomyosarcoma, respectively. Both cases are treated with chemotherapy and show a good response with no evidence of recurrence or metastasis. The aim of this paper is to expand the differential diagnosis of primary mesenchymal kidney tumors in pediatric age group.

1. Introduction

The World Health Organization (WHO) classified rhabdomyosarcoma (RMS) into four major groups, embryonal RMS (ERMS), alveolar RMS (ARMS), pleomorphic RMS (PRMS), and spindle cell/sclerosing RMS (SRMS) [1]. Each one of these entity have its own histopathology criteria for diagnosis [2]. Embryonal RMS commonly arise in head and neck region, genitourinary system as a suddenly enlarging mass. It typically affect infants and young children. While, alveolar RMS frequently arise in deep soft tissue of extremities and trunk, typically in adolescents and young adults, and fellow a worse prognosis than embryonal. Pleomorphic RMS is high-grade sarcoma of adulthood composed of varying numbers of pleomorphic rhabdomyoblasts, most commonly arising in deep soft tissue of lower extremity, particularly thigh. It's an aggressive disease with frequent early metastasis. The last WHO updates classify a spindle cell RMS as a subtype affecting both children and adults that features prominent spindle cell morphology. The current consideration of this variant is to be very closely related to sclerosing RMS and not embryonal RMS. Overall, none of these variants are commonly noticed to raise in the kidney, especially in pediatric age group [2].

2. Case presentation one

A 2-year-old female patient presented with distended abdomen. Physical examination reveals a very irritable child with palpable abdominal mass in the left upper quadrant. Computed tomography (CT) scan of abdomen and pelvis reveals a large retroperitoneal heterogeneous mass arising from the left kidney measuring 10.8 × 8.9 × 9.9 cm, occupying mainly the upper pole of the kidney (Fig. 1A & 1B). The renal vein and artery are patent. The radiology differential diagnosis was Wilms tumor. Metastatic work up was negative. She underwent renal mass biopsy which reveals embryonal rhabdomyosarcoma tumor. The patient received neoadjuvant therapy before nephrectomy was performed. After the completion of clinical and histopathology examination, the patient staged as Group III, stage III, T1b N0 M0, intermediate risk. She completed all cycles of VAC chemotherapy for duration of 40 weeks. The patient is free of local recurrence and distant metastases after 31 months of follow-up.

2.1. Macroscopic findings

The left kidney measured 12 × 10 × 8 cm and weighed 580 g. Cross sections reveal a large ill-defined mass with homogenous cut surface measured 11.5 × 10 × 7.5 cm occupying most of the enlarged kidney. Almost 60% of the mass is necrotic. A rim of kidney tissue is seen at the periphery.

2.2. Light microscopy and immunohistochemistry

The neoplasm composed of sheets of malignant embryonal to polygonal cells with hyperchromatic nuclei in a myxoid stroma (Fig. 1D). Frequent pleomorphic multilobated nuclei with prominent nucleoli containing eosinophilic cytoplasm are seen (Fig. 1E). Atypical

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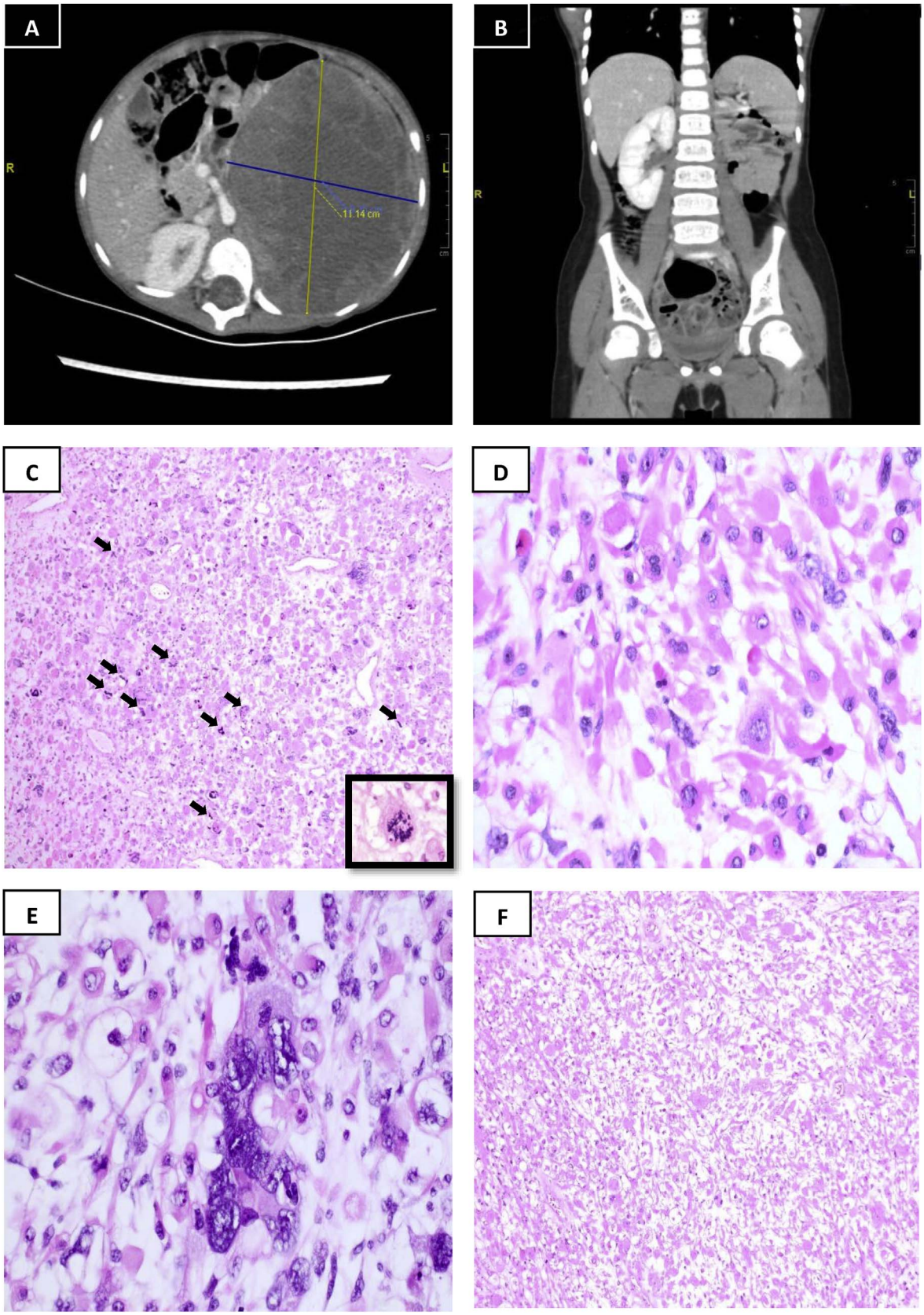

Fig. 1. Case number one; (A) Axial and (B) coronal sections of CT scan of the abdomen showing a well-defined heterogeneous lesion measuring 10.8 × 8.9 × 9.9 cm arising from left kidney; (C) Low-power view showing sheets of malignant cells in a myxoid background, frequent atypical mitosis are seen (arrow head) (H&E; × 10); (D) sheets of embryonal to polygonal cells with eosinophilic cytoplasm and hyperchromatic nuclei (H&E; × 40); (E) Ugly pleomorphic multilobated cell with prominent nucleoli containing eosinophilic cytoplasm (H&E; × 40); (F) Extensive areas of necrosis are seen (H&E; × 10).